5. Management of rheumatic heart disease

The fundamental goal in the long-term management of RHD is to prevent ARF recurrences, and therefore, prevent the progression of RHD, and in many cases allow for the resolution of heart disease.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD).

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to worsening of RHD. Preventing recurrences of ARF by using prophylactic treatment with penicillin is therefore of great importance in controlling RHD.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

Best practice in RHD management

It is difficult and expensive for Aboriginal people and Torres Strait Islanders to travel to major centres for cardiac services, which are often hospital based. Although specialist outreach services are improving in many regions, access to specialist care is suboptimal in rural and remote areas.

The implementation of guidelines for RHD has major implications for Aboriginal and Torres Strait Islander healthcare services, especially in rural and remote regions. In addition to access to appropriate primary care services, best practice for RHD requires:

- secondary prevention with penicillin prophylaxis
- adequate monitoring of anticoagulation therapy in patients with atrial fibrillation and/or mechanical prosthetic valves
- access to oral healthcare
- access to echocardiography
- access to a specialist physician, paediatrician and/or cardiologist, preferably the same specialist, for regular follow up visits
- access to cardiothoracic and interventional cardiology services.

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Valvular lesions in RHD

Specific valvular lesions in chronic RHD include:

- *mitral regurgitation*, in which volume overload of the left ventricle (LV) and left atrium occurs. In more severe cases, this may result in a progressive decline in systolic contractile function
- *mitral stenosis*, where progressive obstruction to left ventricular inflow develops, due to fibrosis and partial fusion of the mitral valve leaflets
- *aortic regurgitation*, where left ventricular volume overload occurs, and there is an increase in left ventricular end-diastolic volume, eventually leading to left ventricular contractile dysfunction in more severe cases
- *aortic stenosis*, which results from fibrosis and fusion of the valve cusps, causing progressive obstruction to left ventricular outflow
- *tricuspid regurgitation*, maybe secondary to left sided rheumatic valve disease or reflect inflammatory rheumatic involvement
- *tricuspid stenosis*, uncommon but causes obstruction to right ventricle inflow (RV).

In patients with multiple valve lesions, management usually focuses on the most severe valve lesion.

Key points in the management of rheumatic mitral regurgitation

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Examination	Pan-systolic murmur at LV apex
Echocardiography	Overriding or prolapse of AMVL Thickened 'dog leg' AMVL, especially if associated with mitral stenosis Retrograde colour (mosaic) regurgitant jet into left atrium, often posteriorly directed Severity graded by area of colour regurgitant jet in left atrium LV chamber dimensions enlarged if moderate or greater MR Assess LV systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	In chronic, stable MR (regardless of severity), there is no role for vasodilators, diuretics or ACE inhibitors unless clinical heart failure is present
Indications for surgery	Moderate / severe MR: 1. NYHA FC II-IV symptoms **OR** 2. Impaired LV systolic function EF <60 % **OR** 3. LVESD ≥ 40 mm in adults or enlarged LVSED Z-score in children **OR** 4. PAS hypertension >50 mmHg **OR** 5. New onset atrial fibrillation
Choice of operation	Mitral valve repair operation of choice Mitral valve replacement with biological or mechanical prosthesis Avoid mechanical prostheses, if concerns about warfarin adherence or future pregnancy

ACE, angiotensin-converting enzyme; AMVL, anterior mitral valve leaflet; EF, ejection fraction; LV, left ventricle; LVESD, left ventricular end-systolic diameter; MR, mitral regurgitation; NYHA FC, New York Heart Association Functional Class; PAS, pulmonary artery systolic.

Key points in the management of rheumatic mitral stenosis

Symptoms	May be asymptomatic Exertional dyspnoea, fatigue, palpitations
Examination	Low-pitched mid diastolic 'rumble' at LV apex
Echocardiography	Thickened restricted 'dog leg' AMVL Restricted posterior leaflet Measure mean mitral diastolic gradient from continuous-wave Doppler signal Calculate MVA from slope of Doppler mitral inflow velocity Calculate PAS pressure
Cardiac catheterisation	Only to exclude coronary artery disease
Atrial fibrillation	Common Rate control using beta-blockers or digoxin Consider cardioversion, if recent onset Need anticoagulation to prevent thromboembolic complications
Medical management	Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic pulmonary venous congestion or pulmonary oedema All symptomatic patients should be referred for cardio-surgical assessment
Indications for intervention	Symptoms NYHA FC II–IV MVA <1.5 cm^2 **OR** PAS pressure >50 mmHg
Procedure of choice	PBMV by high-volume operator/centre Mitral valve repair or replacement if morphology is not suitable for PBMV (e.g. valve is heavily calcified) or if moderate or greater MR is present

AMVL, anterior mitral valve leaflet; LV, left ventricle; MR, mitral regurgitation; MVA, mitral valve area; NYHA FC, New York Heart Association Functional Class; PAS, pulmonary artery systolic; PBMV, percutaneous balloon mitral valvuloplasty.

Key points in the management of rheumatic aortic regurgitation

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Signs	Diastolic blowing and/or decrescendo murmur at left sternal border, usually associated with systolic ejection murmur
Echocardiography	Retrograde diastolic regurgitant colour jet in LVOT and LV chamber Area of jet in LVOT correlates with severity LV chamber dimensions enlarged, if moderate or greater aortic regurgitation Associated mitral valve disease is common Pan-diastolic reversed diastolic flow in descending thoracic aorta, if moderate/severe aortic regurgitation (Doppler) Assess LV systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	All symptomatic patients should be commenced on an ACE inhibitor and referred for cardio-surgical evaluation Consider ACE inhibitors or vasodilator therapy with dihydropyridines (e.g. nifedipine) in asymptomatic patients with moderate or greater aortic regurgitation , especially if systolic hypertension is present
Indications for surgery	Moderate/severe aortic regurgitation with symptoms NYHA FC II–IV Asymptomatic moderate/severe aortic regurgitation if: • LVEF <55% **OR** • LVESD ≥55 mm **OR** • LVEDD >70 mm **OR** • Enlarged LVESD or LVEDD Z-score (in children only)
Choice of surgery	1. Bioprosthetic or homograft valve replacement: • no requirement for anticoagulation if in sinus rhythm • limited durability in younger patients 2. Mechanical valve replacement: • anticoagulation is required 3. Aortic valve repair: • many centres have limited experience 4. Ross procedure (replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft): • only in selected cases with experienced surgeons

ACE, angiotensin-converting enzyme; LV, left ventricle; LVEDD, left ventricular end-diastolic diameter; LVEF, left ventricular ejection fraction; LVESD, left ventricular end-systolic diameter; LVOT, left ventricular outflow tract; NYHA FC, New York Heart Association Functional Class.

Key points in the management of rheumatic aortic stenosis

Symptoms	May be asymptomatic Exertional dyspnoea, angina, syncope
Signs	Low-pitched, systolic ejection murmur in aortic area
Echocardiography	Thickened, restricted aortic valve leaflets Measure peak and mean systolic gradient from Doppler velocity across aortic valve Assess left ventricular systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	Medical therapy is not indicated in asymptomatic patients Symptomatic patients require surgery and do not benefit from medical therapy
Indications for surgery	Symptoms plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm^2 Impaired cardiac function (EF < 50%) plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm^2
Choice of surgery	Bioprosthetic or homograft valve replacement: • limited durability • no requirement for long-term anticoagulation if in sinus rhythm Mechanical valve replacement: • long-term anticoagulation is required

AVA, aortic valve area; EF, ejection fraction.

Key points in the management of rheumatic tricuspid regurgitation

Symptoms	Exertional dyspnoea and fatigue, usually secondary to left sided rheumatic valve disease
Examination	Elevated jugular venous pressure with prominent v wave in jugular pulse Pansystolic murmur left sternal border Hepatomegaly, may be pulsatile Ascites Peripheral oedema
Echocardiography	Thickened leaflets Retrograde colour jet into right atrium Severity graded by area of colour jet Dilated IVC Retrograde flow in hepatic veins Right ventricular chamber enlargement if moderate or greater TR
Medical Management	Symptoms are generally related to the left sided valve lesions Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure Note: Usually impossible to distinguish rheumatic from non-rheumatic tricuspid valve regurgitation clinically or by echocardiogram
Indications for surgery	Moderate/severe TR usually in association with symptomatic MVD Progressive symptomatic right heart failure
Choice of surgery	Tricuspid valvuloplasty Tricuspid valve replacement with mechanical or biological prosthesis if valvuloplasty not possible

IVC, inferior vena cava; MVD, mitral valve disease; TR, tricuspid regurgitation.

Key points in the management of rheumatic tricuspid stenosis

Symptoms	Usually secondary to left sided rheumatic valve disease
Examination	Elevated jugular venous pressure Prominent a wave in jugular pulse Presystolic and mid diastolic murmur at the left sternal border
Echocardiography	Thickened, restricted tricuspid valve leaflets with doming Diastolic gradient measured across tricuspid valve as per MS
Medical management	Symptoms are generally related to the left sided valve lesions Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure
Indications for surgery	Moderate/severe TS in association with symptomatic MVD Progressive right heart failure
Choice of surgery	Percutaneous balloon valvuloplasty or surgical commisurotomy operation of choice Tricuspid valve replacement with mechanical or biological prosthesis if repair or PBTV not possible

MS, mitral stenosis; MVD, mitral valve disease; PBTV, percutaneous balloon tricuspid valvuloplasty; TS, tricuspid stenosis.

RHD Australia

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The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides include:

- Primary prevention of ARF
- Diagnosis of ARF
- Management of ARF
- Secondary prevention of ARF
- Management of RHD
- RHD in pregnancy
- RHD control programs

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